460* Gender differences in the Scandinavian CF population

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Aims: Poorer survival, earlier acquisition of *Pseudomonas aeruginosa* (P.a.) and higher prevalence of diabetes mellitus (DM) for female CF patients have been reported. The aim of this study was to see if this was also true for CF females in Scandinavia.
Methods: Data from the Scandinavian Pseudomonas Prevalence Study were analysed for gender differences. 898 (420 F/478 M) pancreatic insufficient patients were included.
Inclusion data: age, BMI, lung function, bacterial colonisation, DM. Chronic P.a. infection was defined as serum precipitins of $\geqslant 2$ at inclusion and end of study (14 months). Days of intravenous antibiotics and hospitalisation were recorded.
Results: 195 females (46%) and 195 (40%) males had a chronic P.a. infection ($p = 0.089$). Females had higher prevalence in all 5-year age-groups, though only significantly in the 30+ group ($p = 0.031$). 16 females (4%) and 9 males (2%) had Burkh. infection ($p = 0.067$). Adult women had a median of 39 days on i.v. antibiotics/year versus 26 days for men ($p = 0.01$), and spent more days in hospital (mean 20 versus 13; $p = 0.01$). No difference was seen in lung function and BMI. 60 females (14%) had DM versus 59 males (12%), $p = 0.43$. 12 females (3%) and 6 males (1%) died or were lung transplanted during the study ($p = 0.087$). Mean age of death/lung transplantation was 27 y for females and 22 y for males ($p = 0.26$). Significantly more adult males (56%) than females (44%) implied difference in mortality.
Conclusions: A trend was seen towards a higher prevalence of chronic P.a. infections in females as well as more days of i.v. antibiotics and hospitalisation. Since the adult group had more males than females, the lack of differences in lung function, BMI and diabetes might be due to an excess mortality in the female patients, hinted in the doubled rate of death/lung transplantation among the women.

461* Is CF more severe in females (F) than in males (M)? – Survival analysis from the Italian Cystic Fibrosis Registry (ICFR)

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According to some, but not all, epidemiological studies, CF is more severe and causes earlier mortality in females (F) than in males (M).
Methods: the Italian Cystic Fibrosis Registry (ICFR) was implemented in 1988 and 30 CF Italian centres prospectively collected data of patients alive on Jan 1st, 1988 and of all those diagnosed thereafter ($n = 5191$). We performed survival analysis on 5175 subjects (F 2522) according to gender, period and mode of diagnosis.
Results: 717 deaths were related to CF (F 385, M 332; $p < 0.009$). Kaplan–Meier survival curves were highly different between F and M (Log-rank $p < 0.0006$). Survival probability at age 18 years was 90% for F and 92% for M. The data were period dependent: M and F born $\geqslant$1988 (M 1183, F 1170; 78 deaths) had not significantly different survival, either those diagnosed by neonatal screening ($n = 512$) or by symptoms ($n = 928$). Moreover, while numbers of F and M are equal for patients born $\geqslant$1988, the proportion of M was higher among those born <1988. This could be a selection bias due to higher precocious mortality of F. A similar number of F and M underwent lung transplantation.
Conclusions: taken together, these data indicate that earlier death in F should no longer be seen in the more recent cohorts of patients. Analysis of lung function data in the registry is ongoing to determine whether or not lung disease is more severe in F although not causing earlier mortality.
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462 The Cystic Fibrosis Siblings Study

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Parents of CF children face difficult decisions concerning future pregnancies. For some, antenatal diagnosis and termination of a second CF child is undesirable. This study was designed to determine whether having siblings with CF adversely affects clinical outcome. We hypothesised that those with CF siblings do less well because of factors such as cross infection.
Method: The South and West CF Database was used to identify the following; those with no CF affected sibling, those with one or more affected CF siblings (including CF twins). A dataset from 1990 to 2004 was analysed, thus providing a series of cross-sectional analyses. Outcome measures were compared using t and Mann-Whitney U tests where appropriate.
Results: Three cohorts were analysed; 284 children aged 7 years (232 without CF siblings, 52 with CF siblings, 14 twins), 299 aged 10 years (241 without CF siblings, 58 with CF siblings, 14 twins), 270 aged 15 years (200 without CF siblings, 70 with CF siblings, 5 twins). In all cohorts there was a significant difference for growth of Pseudomonas in the last year, (with CF sibling 86/175 vs without CF sibling 332/673, $p < 0.01$, CF twins 21/33 vs without CF sibling 332/673, $p < 0.01$). There was no significant difference for other outcome measures at any age and after analysing outcomes for siblings separately. (FEV1 87% vs 85%, banding score 2.16 vs 2.2, BMI SDS –0.07 vs –0.04 for aggregated data).
Conclusion: Despite differences in Pseudomonas colonisation there were no differences in clinically relevant outcomes indicating more severe disease. These data provide useful information for parents of CF children considering future pregnancies. In this study we found no evidence that children with CF are adversely affected when there are two or more siblings in the same family.

463* Improved survival in Cystic Fibrosis associated with the specialized center care

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To improve the survival and quality of life of cystic fibrosis (CF) patients in Republic of Macedonia, the program for development of specialized CF center at the Pediatric Clinic in Skopje was established in 1996/97.
The aim of the study was to evaluate the results of the new therapeutic approaches and management of CF patients, by analyzing of the demographic data (age, gender, age of death if applicable) and the cumulative survival of CF patients treated at our clinic over two periods: from 1989 to the end of 1996, and from 1997 to the end of 2004.
The First 8 years, 63 CF patients (37 males, 26 females) were treated. A great number of those patients (28 or 44.4%) died during that period, most of them (21 or 75%) in infancy. The mean age of death was 1.83 ± 2.73 years. The mean age of survived patient at the end of the firs study period was 6.48 ± 3.81 years. The second 8 years, 94 CF patients (57 males, 37 females) have attended the CF centre. During that period 6 (6.38%) deaths occurred, 2 (33.3%) of them in young infant age. The mean age of death was 4.04 ± 4.11 years. The mean age of alive patients at the end of the second period was 9.55 ± 6.14 years and they were significantly older than the patients in the first period. CF population in Macedonia is getting larger and older. The proportion of the patients older than 12 years (13 years and more) at the end of 2004 was 32.9% in comparison with only 5.7%, at the end of 1996. The estimation of cumulative survival in the two analyzed periods showed significantly higher cumulative survival in the second period (0.908) compared to first period (0.455).
In conclusion, the lifetime provision of medical care within a specialized CF centre is associated with considerable decrease of mortality rate and improved survival of CF patients.